

Leukaemia Section

Short Communication

t(3;7)(q27;p12-13)

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Clinics and pathology

Disease

Non Hodgkin lymphoma (NHL).

Phenotype / cell stem origin

2 cases of diffuse large B-cell lymphoma (DLBL) with BCL6/ZNFN1A1 involvement, 1 NHL with BCL6 involvement, 1 follicular, predominantly large cell B-NHL and one T-NHL without gene ascertainment.

Epidemiology

5 cases to date, aged 40-70 years, sex ration: 3M:1F.

Prognosis

The 2 patients with BCL6/ZNFN1A1 involvement died at 16 and 17 months after diagnosis.

Cytogenetics

Additional anomalies

del(6) in two cases, del(13q) in two cases, +16 in two cases, +12 in one case.

Genes involved and Proteins

BCL6

Location: 3q27

Protein

Transcription factor; belongs to the Krüppel family, with a N-term BTB/POZ domain and 6 zinc fingers; transcription repressor.

ZNFN1A1 (Ikaros)

Location: 7p12

Protein

Transcription regulator; can repress transcription through the recruitment of histone deacetylase complexes; role in conjunction with Aiolos; hemopoietic-specific zinc finger protein regulator of B and T-cell differentiation.

Results of the chromosomal anomaly

Hybrid gene

Description

5' Ikaros - 3' BCL6 fusion transcript; it is supposed that substitution of the promoter of BCL6 may be responsible for BCL6 deregulation.

References

Konishi H, Sakurai M, Nakao H, Maseki N, Kaneko Y, Yagiri Y, Notohara K, Frizzera G. Chromosome abnormalities in malignant lymphoma in patients from Kurashiki: histological and immunophenotypic correlations. *Cancer Res* 1990;50:2698-2703.

Bastard C, Deweindt C, Kerckaert JP, Lenormand B, Rossi A, Pezzella F, Fruchart C, Duval C, Monconduit M, Tilly H. LAZ3 rearrangements in non-Hodgkin's lymphoma: correlation with histology, immunophenotype, karyotype, and clinical outcome in 217 patients. *Blood* 1994;83:2423-2427.

Brown KE, Guest SS, Smale ST, Hahm K, Merkenschlager M, Fisher AG. Association of transcriptionally silent genes with Ikaros complexes at centromeric heterochromatin. *Cell* 1997;91:845-854.

Georgopoulos K, Winandy S, Avitahl N. The role of the Ikaros gene in lymphocyte development and homeostasis. *Annu Rev Immunol* 1997;15:155-176.

Nichogiannopoulou A, Trevisan M, Friedrich C, Georgopoulos K. Ikaros in hemopoietic lineage determination and homeostasis. *Semin Immunol* 1998;10:119-125.

Ichinohasama R, Miura I, Funato T, Sato I, Suzuki C, Saito Y, Decoteau JF, Myers JB, Kadin ME, Sawai T, Ooya K. A recurrent nonrandom translocation (3;7)(q27;p12) associated with BCL-6 gene rearrangement in B-cell diffuse large cell lymphoma. *Cancer Genet Cytogenet* 1998;104:19-27.

Tanaka K, Arif M, Eguchi M, Guo SX, Hayashi Y, Asaoku H, Kyo T, Dohy H, Kamada N. Frequent allelic loss of the RB, D13S319 and D13S25 locus in myeloid malignancies with deletion/translocation at 13q14 of chromosome 13, but not in lymphoid malignancies. *Leukemia* 1999;13:1367-1373.

Hosokawa Y, Maeda Y, Ichinohasama R, Miura I, Taniwaki M, Seto M. The Ikaros gene, a central regulator of lymphoid differentiation, fuses to the BCL6 gene as a result of t(3;7)(q27;p12) translocation in a patient with diffuse large B-cell lymphoma. *Blood* 2000;95:2719-2721.

Payne KJ, Huang G, Sahakian E, Zhu JY, Barteneva NS, Barsky LW, Payne MA, Crooks GM. Ikaros isoform x is selectively expressed in myeloid differentiation. *J Immunol* 2003;170:3091-3098.

Liberg D, Smale ST, Merckenschlager M. Upstream of Ikaros. *Trends Immunol* 2003;24:567-570.

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